reversible encephalopathy syndrome within 50 days post transplantation, all of them showed complete clinical and radiological resolution. **Conclusion:** Non-myeloablative HSCT using this conditioning regimen for high risk pediatric patients with benign hematological disorders appears to be promising and worth further study.

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PERICARDIAL EFFUSION (PEF) IS A MAJOR INDEPENDENT RISK FACTOR ASSOCIATED WITH A SIGNIFICANT DECREASE IN SURVIVAL IN PEDIATRIC STEM CELL TRANSPLANTATION (SCT) RECIPIENTS

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Background: SCT is the treatment of choice for a variety of pediatric conditions. Potential post SCT morbidities include cardiac toxicity. Rhodes et al (BMT 2005) reported that PEF was seen in 4.4% of pediatric SCT recipients. Risk factors for the development of PEF have been thought to include GVHD, infection and/or underlying disease. There is a paucity of information regarding the etiology, prognosis, treatment and outcome of PEF in a large cohort of pediatric SCT recipients. **Objectives:** To assess the incidence, risk factors, outcome of PEF and the impact of PEF on overall survival (OS) in pediatric SCT recipients. **Method:** Echocardiograms were performed at baseline prior to 200 SCT (4 were ineligible for review) in 156 patients and when patients had symptoms and/or signs of cardiac or pericardial disease post SCT. Probability of and time to PEF were analyzed by Kaplan-Meier method and risk of PEF and death were determined by multivariate analysis. Covariates included age, gender, ethnicity, conditioning regimen, risk status (CIBMTR criteria), conditioning regimen, donor source, CMV status, GVHD, and HLA match. **Results:** The mean age was 8.15 years (+/- 6.25 years) with 88 males and 68 females. 102 patients received allogeneic transplants, 34 of them received more than 1 transplant. 116 of 156 recipients had malignant disease. 100 of 156 patients had ablative conditioning. The incidence of PEF was 14.7%. A multivariate analysis shows that older age, poor risk and unrelated cord blood donor recipients are significantly associated with a risk of developing PEF with hazard ratios of 1.125 (1.051–1.205), 3.508 (1.502–8.191), and 5.080 (1.141–22.625), respectively. OS was significantly decreased in patients with PEF versus without PEF (hazard ratio = 4.84, 95% CI, 2.814–8.322, $p < 0.0001$). Furthermore, in a multivariate analysis, there was a significant decrease in OS associated with PEF, CMV status and poor risk status with hazard ratios of 3.296 (1.73–6.26), 1.89 (1.08–3.30) and 1.93 (1.12–3.3), respectively. **Conclusion:** These results demonstrate an almost 15% incidence of PEF in pediatric SCT recipients. Older, poor risk and UCB donor recipients may be at higher risk of developing PEF. PEF was associated with the most significant impact on overall mortality independent of other risk factors. Improved prevention and therapeutic strategies for development of PEF in post allo SCT recipients may potentially reduce mortality in the future.

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HEMATOPOIETIC STEM CELL TRANSPLANTATION (HSCT) IN WISKOTT ALDRICH SYNDROME (WAS): A SINGLE CENTER EXPERIENCE IN 25 PATIENTS

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WAS is a rare X-linked disease characterized by eczema, microthrombocytopenia, recurrent infections, autoimmune complications and malignancies. The gene responsible for WAS has been identified and termed *WAS*P. Without a successful HSCT, the prognosis of classical WAS with a complete absence of WASP expression is poor. In this report we analyze the outcome of 25 pts transplanted in a single center in a developing country.

Material and Methods: Period: 04/92–02/07; age:1–14 ys-old(M:2 ys). Eight pts received bone marrow(BM) from HLA matched (5 pts) or mismatched family donors(3 pts). 17 pts received an unrelated cord blood(URD CB). Preparatory regimen: Busulfan(BU)16 mg/kg+Cyclophosphamide 120–200 mg/kg in 24 pts or BU8 mg/kg + Fludarabine 125 mg/m^2:1 pt. ATG(rabbit) was added to patients transplanted from unrelated donors or mismatched family donors. GVHD prophylaxis: Cyclosporine(Csa)+ MTX:16 pts; Csa + steroids:8 pts and Csa + MMF:1 pt. After 2005, all pts submitted to an URD CB transplantation, received Csa+MTX as GVHD prophylaxis if the TNC infused was $> 3.0 \times 10^*7$/kg. **Results:** 21 pts(84%) are alive between 100 – 4745 days (M:790 d) after HSCT. All pts survived more than 28 days and were evaluable for engraftment. Primary graft failure (PGF) occurred in 2 pts (both URD CB 4/6) and one pt was submitted to a successful 2nd transplant. Most pts tolerated the procedure very well. See table below for transplant related complications.23/25 pts were analyzed for chimerism (VNTR). 13 pts had a complete chimerism. Ten pts were mixed chimeras immediately after HSCT and, later on, 3 pts became full chimeras. Four pts had autoimmune complications after transplant: 3 pts had idiopathic thrombocytopenic purpura(ITP) and one pt had severe auto immune hemolytic anemia (AIHA). Frequent viral infections (CMV, EBV, adenovirus) occurred in pts submitted to URD CB transplants. 2/21 surviving pts have permanent sequels from neurological viral infections. **Conclusions:** The excellent survival in this group of pts confirms the efficacy of HSCT in this disease. Pts receiving HSCT from 4/6 URD CB have a lower survival. Reactivation of viral infections were frequent complications in our pts and should be monitored closely.

Post-transplant complications

	URD CB Compatibility: 6/6 or 5/6 N = 11 pts	URD CB Compatibility: 4/6 N = 6 pts	BM from matched family donors N = 5 pts	BM from mismatched family donors N = 3 pts
Graft failure	0	2	0	0
Acute GVHD grade III-IV	3/11	1/4	0	0
Chronic GVHD extensive	2/11	1/4	0	0
Mixed chimeras	3 pts	2 pts	2 pts	0
Auto immune complications	0	2 pts (ITP and AIHA)	1 pt (ITP)	1 pt (ITP)
Overall Survival	91%	66%	80%	100%
Causes of death	1 pt: D + 1825: bronchopneumonia + respiratory failure	2 pts: D + 34 : PGF+ fungal infection and D + 132 : C-GVHD + bacterial sepsis	1 pt:D + 62: CMV pneumonia (1992)	0

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IRON OVERLOAD IN PEDIATRIC HEMATOPOIETIC STEM CELL TRANSPLANT (HSCT) PATIENTS

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Background: Iron can be a highly toxic molecule to cells and tissues when it interacts with oxygen and generates free radicals. Free iron, ferritin and transferrin saturation have been shown to increase acutely in HSCT patients, and high ferritin levels can persist for years following HSCT. The prevalence of iron overload has not been defined in this population, and currently, no management guidelines exist. We describe a group of pediatric HSCT patients who were diagnosed with iron overload after developing liver function abnormalities and/or hyper-ferritinemia. **Methods:** We performed a detailed retrospective review of patients identified as having iron overload following allogeneic HSCT performed between 2001–2007. We included a subset of patients with known